ABBREVIATIONS USED IN TEXT

CNS = central nervous system
CSF = cerebrospinal fluid
CT = computed tomography
EEG = electroencephalogram

ment with acute meningoencephalitis, organic psychosis or progressive dementia; (2) predominantly brain-stem or spinal cord involvement with cranial nerve palsies, hemiparesis, paraparesis, ataxia, pseudobulbar palsy or parkinsonism, and (3) thrombosis of major intracranial draining veins causing intracranial hypertension with headaches and papilledema. Once CNS involvement is present, a relapsing and fluctuating course develops in about half the patients, with permanent neurologic sequelae. Fortunately, some patients improve remarkably, as did our patient who recovered from several bouts of delirium and two episodes of coma.

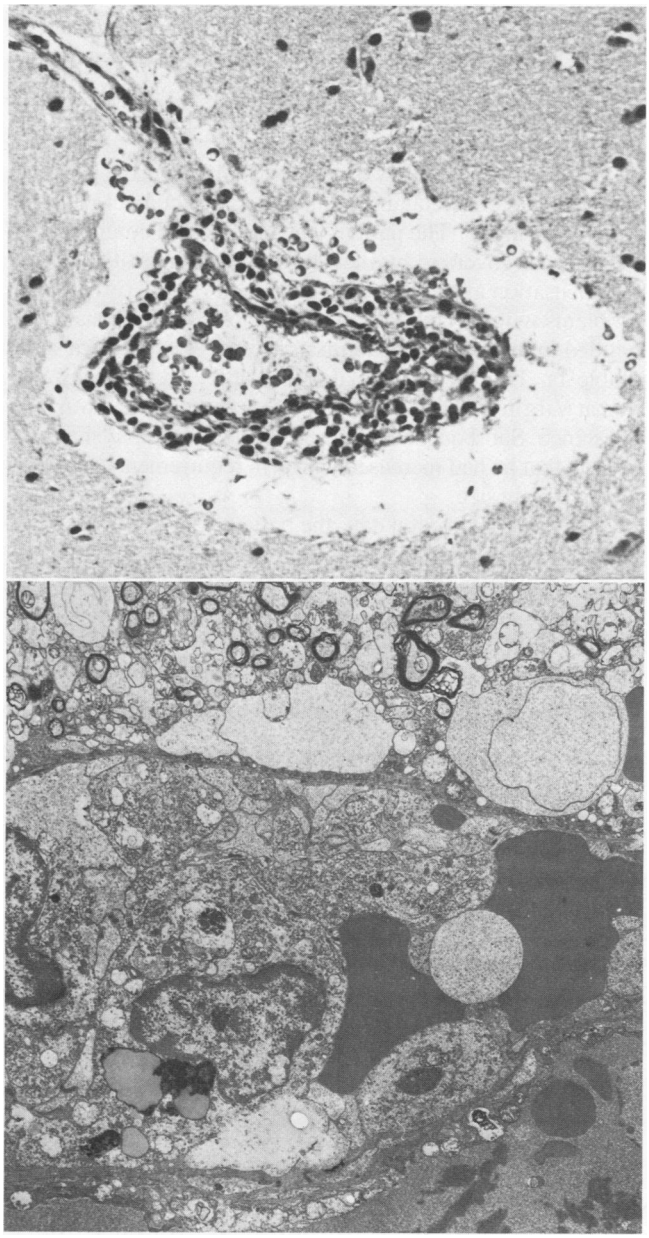

Figure 1.—Photomicrographs of right frontal cortex. **Top**, The perivascular space contains numerous large and small mononuclear cells; its outer limit is separated by artifact from surrounding parenchyma, which is histologically normal (hematoxylin and eosin, × 190). **Bottom**, Electron micrograph shows aggregated macrophages distending the perivascular space of a small blood vessel cut longitudinally. A few extravasated erythrocytes are admixed (× 11,900).

Behçet's syndrome should be included in the differential diagnosis of unexplained recurrent meningoencephalitis. A history of recurrent aphthous ulcers of the lips and genital areas, iritis and other signs of Behçet's disease should be sought. Although the disease is most common in Middle Eastern and Japanese men, Behcet's disease does occur in the United States.

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Münchausen's Syndrome Presenting With Postphlebitic Syndrome

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MÜNCHAUSEN'S SYNDROME is feigned illness presented in a plausible manner to gain hospital admission. The illness is listed as a chronic factitious disorder with physical symptoms in the American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders.*[1] The syndrome takes its name from Baron Karl Friedrich Hieronymus von Münchhausen.[2] The 18th-century German baron was known to entertain friends with greatly exaggerated stories. Asher, in 1951, first used the name Münchausen to describe three patients with dramatic gastrointestinal complaints that could not be correlated with surgical or pathologic findings.[3] In recent years, the syndrome has described hospital abusers with con-

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cealed histories of extensive travel and hospital admissions. Patients frequently have self-inflicted signs and symptoms, undergo elaborate medical and surgical investigations and leave hospitals against medical advice.[4]

Patients have targeted all organ systems for their fictional symptoms. In the following report, we describe a patient with the postphlebitic syndrome posing as a case of recurrent acute deep vein thrombophlebitis.

Report of a Case

The patient, a 46-year-old morbidly obese man, presented in February 1984 to the Phoenix Baptist Hospital emergency room with a five-day history of bilateral leg pain and paresthesias and swelling of his left calf.

The patient's home was in Montana, but he was itinerant and had no local physician. He reported having recurrent thrombophlebitis, possibly 30 episodes, and several episodes of pulmonary embolus. His original injury was reported as a gunshot wound to his left lower leg in 1979. He was then employed as an industrial security guard and all related medical care was covered by Workers' Compensation. His first episode of thrombophlebitis followed soon after. An inferior vena caval umbrella was placed in 1981, following his second pulmonary embolus, confirmed by findings of a lung scan. He reported three more episodes of pulmonary embolus after that operation, however. His last episode of thrombophlebitis occurred two months earlier, at a hospital in Albuquerque he could not remember. The patient reported a "bad venogram" was done at that time and he was admitted to hospital for two weeks of intravenous heparin therapy.

The patient said he had not had trauma, long car rides or other prolonged periods of leg inactivity during the previous two weeks. He stated that his current symptoms exactly mimicked his previous episodes of thrombophlebitis. His physician in Montana, he said, recommended hospital admission for intravenous heparin therapy whenever he experienced these symptoms.

The patient said that resistance to warfarin sodium (Coumadin) developed. He reported being studied at the University of Washington Hematology Section in Seattle. They recommended a daily warfarin dose of 50 mg. The patient had several bleeding episodes on that dosage, however, and lowered it himself.

He reported compliance with his only current medication of 20 mg of warfarin daily. His family history was positive for exogenous obesity in both parents and all siblings. The patient had smoked one pack of cigarettes a day for the past seven years and occasionally used alcohol.

On physical examination he weighed 133 kg (294 lb) and complained of left leg discomfort. The patient was afebrile and other vital signs were normal. His chest was clear bilaterally and heart sounds were muffled by his large chest. The abdomen was soft and nontender. The left lower leg was the most tender, but mild tenderness was noted on palpation of the entire lower extremities. Minimal erythema was observed in the lower part of the left leg. Lower leg circumferences for both legs were equal when measured 5 cm below the tibial tubercles. The patient grimaced on Homan's testing of the left leg. Moderate edema was present at the ankles bilaterally, improving with elevation. There was hyperpigmentation of the skin, decreased hair growth and mild atrophy of the skin over the medial lower legs. No ulcerations or scarring were present.

Studies done on admission showed normal thyroid function, chest film and electrocardiogram, and results of chemistry tests and urinalysis were normal. The leukocyte count was 13,300 per μl with a normal differential. The triglyceride level was elevated to 435 mg per dl. Prothrombin time was 14.7 seconds with a control of 11.6 seconds. The partial thromboplastin time was normal.

The patient was admitted to hospital to rule out recurrent thrombophlebitis. He was given a 10,000-unit bolus of heparin intravenously, followed by 1,500 units an hour by continuous intravenous drip. His left leg was elevated and heat applied. The next morning, a vascular surgeon was consulted. The diagnosis of postphlebitic syndrome was confirmed from the constellation of chronic phlebitis changes evident in the legs. A venogram was not done since it was believed that one was abnormal in the recent past.

Several phone calls were made to obtain further history. The patient's physician in Montana had lost contact with him two years earlier. The patient was granted full compensation for medical coverage by the Montana Division of Workers' Compensation after being hit by a gate in 1980. His recurrent problems with thrombophlebitis and possible pulmonary emboli led to many hospital admissions in several western states (Table 1). Those who evaluated him at the Hematology Section at Virginia Mason Hospital in Seattle felt he had warfarin resistance. Subsequent bleeding episodes led his physicians to believe that he had increased warfarin requirements due to his massive body size.

On the third hospital day, the patient was receiving 2,000 units of heparin per hour, as well as 20 mg of warfarin daily. The partial thromboplastin time was 66.4 seconds with a control of 30.7 seconds. The prothrombin time was 18.0 seconds with a control of 11.9 seconds. The patient's symptoms persisted and he became very demanding of nursing personnel. He became vague in his historical data as he was confronted with previously concealed information. Rapid resolution of his symptoms was noted on the fourth day.

The diagnosis of Münchausen's syndrome had been entertained during several previous hospital admissions. Psychiatric consultation, obtained during this admission, confirmed the diagnosis of a fictional disorder consistent with Münchausen's syndrome.

The patient was discharged on the fifth hospital day for outpatient follow-up. The daily warfarin dose was continued at 20 mg. The patient refused to wear support stockings or to quit smoking, as recommended. He did not keep his follow-up outpatient appointments.

Discussion

The postphlebitic syndrome is a recognized complication of deep vein thrombosis. The syndrome is estimated to exist in 1% to 2% of the general population.[5] In accordance with the incidence of deep vein thrombosis, the syndrome is more common in the left leg.

The loss of valvular competence in the deep venous system produces ambulatory venous hypertension and corresponding skin changes.[6] Ulceration has been reported in as many as 20% of patients within five years of the onset of the disease. Stasis pigmentation, alopecia and edema were all seen in this

patient. Pain has also been frequently recognized as a component of this syndrome.[6] The rapid disappearance of this patient's pain after he was confronted with the concealed medical history made his account of pain suspect.

The clinical diagnosis of deep vein thrombosis is notoriously inaccurate.[7] The disease is frequently present in the absence of symptoms, as well as being absent in half of patients in whom clinical signs or symptoms suggest its presence. The radiologic standard for identifying deep vein thrombosis is the venogram. The age of the thrombotic changes is not always apparent from the venogram,[8] so the test may not be able to distinguish new from old disease. With the postphlebitic syndrome, the deep venous system may show residual abnormalities. Chronic disease frequently causes the contrast medium to flow preferentially through the superficial system.[8]

Chronic venous change may also make it technically difficult to inject contrast medium into the leg for a venogram. Two noninvasive procedures used in detecting disease include impedance plethysmography and Doppler flow studies. Radiofibrinogen leg scanning is also used, but this test also requires injection. While these tests all have their limitations, they can greatly aid in diagnosis when selectively done together or in combination with the venogram.[7] Several newer tests may be helpful in diagnosing chronic disease and the postphlebitic syndrome. Descending (rather than ascending) venography has shown efficacy in diagnosing chronic venous damage.[9] Elevated Doppler venous pressures have been noted with the postphlebitic syndrome.[10]

TABLE 1.—*List of Hospital Admissions**

October 1980	Deaconess Hospital	Billings, Mont
November 1980	St Vincent's Hospital	Billings, Mont
November 1980	Deaconess Hospital	Billings, Mont
December 1980	St Vincent's Hospital	Billings, Mont
December 1980	Central Montana Hospital	Lewistown, Mont
February 1981	St Patrick's Hospital	Missoula, Mont
March 1981	Deaconess Hospital	Great Falls, Mont
March 1981	St Patrick's Hospital	Missoula, Mont
April 1981	Kalispell Regional Hospital	Kalispell, Mont
July 1981	St James' Hospital	Butte, Mont
September 1981	Penrose Hospital	Colorado Springs, Colo
September 1981	Memorial Hospital	Colorado Springs, Colo
October 1981	McKee Medical Center	Loveland, Colo
October 1981	Sheridan County Memorial Hospital	Sheridan, Wyo
November 1981	St James' Hospital	Butte, Mont
November 1981	Virginia Mason Hospital	Seattle
December 1981	St James' Hospital	Butte, Mont
January 1982	Desert Springs Hospital	Las Vegas
March 1982	Virginia Mason Hospital	Seattle
April 1982	St Peter's Hospital	Helena, Mont
July 1982	St Patrick's Hospital	Missoula, Mont
October 1982	St Patrick's Hospital	Missoula, Mont
November 1982	Coulee General Hospital	Grand Coulee, Wash
June 1983	St Peter's Hospital	Helena, Mont
July 1983	St Peter's Hospital	Helena, Mont
October 1983	St Peter's Hospital	Helena, Mont
November 1983	St Peter's Hospital	Helena, Mont
November 1983	Desert Springs Hospital	Las Vegas
December 1983	Desert Springs Hospital	Las Vegas
December 1983	Heights General Hospital	Albuquerque
January 1984	Scottsdale Memorial Hospital	Scottsdale, Ariz
January 1984	St Luke's Hospital	Phoenix
February 1984	Phoenix Baptist Hospital	Phoenix
February 1984	Phoenix General Hospital	Phoenix
March 1984	Desert Springs Hospital	Las Vegas
April 1984	Southern Nevada Memorial Hospital	Las Vegas
May 1984	Desert Springs Hospital	Las Vegas
September 1984	Seward General Hospital	Seward, Alaska

*Compiled in December 1984.

Patients presenting with possible thrombophlebitis generally should undergo diagnostic confirmation before instituting heparin therapy.[7] This patient presented in the evening and heparin therapy was begun before study, until his suspicious history and findings could be scrutinized. The morning following admission, it was learned that numerous studies during several previous hospital admissions revealed chronic disease. A venogram done just several weeks before this admission, in New Mexico, showed only chronic changes.

Patients with Münchausen's syndrome frequently submit to excessive medical testing. Repeated diagnostic testing and medical attention may support the unusual behaviors exhibited by Münchausen's patients. In light of the recently confirmed chronic disease and good support for the diagnosis of Münchausen's syndrome, our consultants suggested minimal further diagnostic workup.

Many of the salient features of chronic factitious illness were evidenced in this patient. His illness began with a legitimate physical problem. The patient had a concealed history of many hospital admissions with extensive travel. His medical history had a dramatic flair initially; however, the patient became vaguer and more inconsistent on further questioning. He became very demanding of medical personnel. While the patient did not leave against medical advice, as is common for patients with Münchausen's syndrome, the treatment recommendations were not followed.

The list of this patient's hospital admissions is impressive (Table 1). We were not aware of his many hospital admissions in Phoenix until well after discharge. He visited several hospitals in the same city on different occasions, and he was known to leave against medical advice at least once in 1981.

The charges for hospital admissions and professional fees for Münchausen's patients frequently are not recovered, and the costs are absorbed. This case represented an estimated expense to the state of nearly $100,000. We reported our findings to the Montana Division of Workers' Compensation and recommended reevaluation of this patient's current status.

The diagnosis of Münchausen's syndrome is not easily made in the emergency department or on initial presentation. A suspicion of fictional illness, when coupled with an unusual or inconsistent history and the postphlebitic syndrome, may aid an admitting physician to avoid unnecessary procedures or hospital admission.

The postphlebitic syndrome is not listed as a common manifestation of Münchausen's syndrome.[1,4] We hope this report will encourage physicians to consider this disorder's potential for being used fictitiously.

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Severe Pulmonary Hypertension in a Patient With Rheumatoid Arthritis—Response to Nifedipine

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SEVERE PULMONARY HYPERTENSION in patients with rheumatoid arthritis is rare[1] and is more commonly associated with other collagen-vascular diseases such as progressive systemic sclerosis[1,2] or the syndrome of calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly and telangiectasia.[3] Precapillary pulmonary hypertension in patients with rheumatoid arthritis may be found when the condition is complicated by pulmonary fibrosis.[4] Severe pulmonary hypertension associated with rheumatoid arthritis in those patients with no clinical evidence of pulmonary parenchymal involvement is thought to be caused by pulmonary arteriolar vasospasm or pulmonary vasculitis.[5] This latter group of patients with no apparent parenchymal involvement appears clinically similar to patients with primary pulmonary hypertension except for the arthritis.[1,5,6]

Recent reports of the efficacy of vasodilator therapy in patients with primary pulmonary hypertension[7-10] prompted us to evaluate the calcium channel blocker nifedipine in a patient with rheumatoid arthritis, Raynaud's phenomenon and severe precapillary pulmonary hypertension.

Report of a Case

The patient, a 61-year-old man with a 30-year history of rheumatoid arthritis, presented with dyspnea, episodes of near syncope and Raynaud's phenomenon. He had a 30-pack-year smoking history and had worked in a foundry. For the two years before presentation he was treated with gold and naproxen.

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On physical examination he had normal breath sounds, a right ventricular heave, an increased pulmonic second sound, subcutaneous nodules and joint changes consistent with rheumatoid arthritis. The rheumatoid factor was 1:5,120 and an antinuclear antibody test was negative. An electrocardiogram (Figure 1) showed right atrial enlargement and right ventricular hypertrophy. A chest roentgenogram (Figure 2) showed enlarged main pulmonary arteries and cardiac silhouette. The lung fields were normal.

Pulmonary function data over a 28-month period are listed in Table 1. These values give no evidence of a significant obstructive or restrictive defect. The single breath test of the diffusing capacity of the lung for carbon monoxide and results of a ventilation-perfusion lung scan were normal. There was no activity over the kidneys to suggest a right to left shunt. A two-dimensional echocardiogram showed an enlarged right atrium and right ventricle with paradoxical motion of the

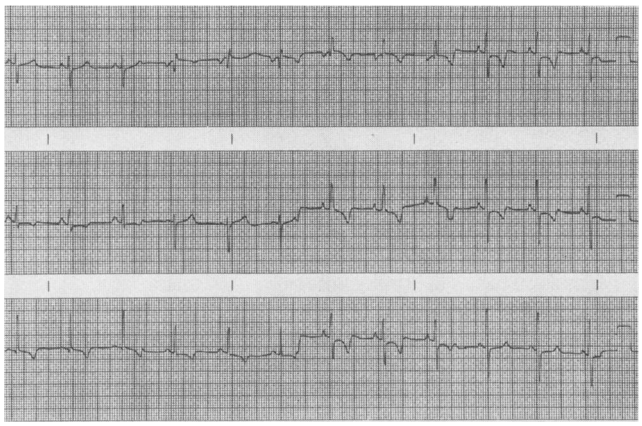

Figure 1.—An electrocardiogram done on a 61-year-old patient shows right atrial enlargement and right ventricular hypertrophy.

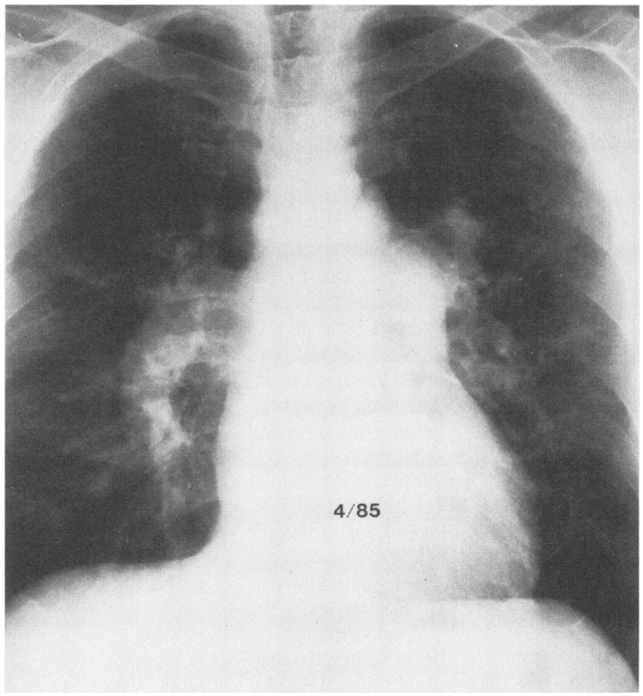

Figure 2.—A chest roentgenogram shows enlarged main pulmonary arteries and cardiac silhouette.